Tension pneumocephalus associated with rupture of a middle fossa encephalocele

Case report

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Acquired nontraumatic (spontaneous) encephaloceles of the middle fossa are not common. Rupture of an encephalocele frequently leads to a cerebrospinal fluid fistula. Tension pneumocephalus consequent to rupture of an encephalocele has not been previously reported, but conceivably occurs by means of a ball-valve mechanism in the ensuing fistulous tract. An unusual case is presented of an elderly man who suffered acute life-threatening neurological symptoms from a tension pneumocephalus that likely developed from rupture of an acquired nontraumatic encephalocele of the left middle fossa. The symptoms correlated with the location of the intracranial abnormalities. The literature is reviewed and the pathophysiology of the lesion is discussed.

Key Words • encephalocele • pneumocephalus • tension pneumocephalus • middle fossa

Acquired nontraumatic (spontaneous) encephaloceles of the middle fossa occur only rarely; until 1990, only six cases had been reported. This type of encephalocele probably occurs where congenital or acquired cranial defects and attenuated dura are present. As with the traumatic form, acquired nontraumatic encephaloceles typically present with headache or evidence of cerebrospinal fluid (CSF) fistulae such as otorhinorrhea and recurrent meningitis, although seizures have also been noted.

Tension pneumocephalus refers to pathological intracranial air under increased pressure. It can arise whenever a ball-valve mechanism exists within an aberrant intracranial air cavity communication. The intracranial contents act as a one-way valve, allowing air to enter into but not escape from the intracranial compartment. Symptoms and signs, such as headache, nausea, vomiting, and a decreased level of consciousness, frequently reflect an increase in intracranial pressure (ICP).

An unusual case of tension pneumocephalus associated with rupture of an acquired nontraumatic encephalocele of the left middle fossa is reported. The dramatic finding of extensive pneumocephalus and focal neurological deficit is an atypical presentation for a patient with encephalocele rupture, although the latter event was not entirely unexpected considering the location of the intracranial air.

Case Report

This 78-year-old man was admitted to another hospital with a 2-day history of progressive confusion, anxiety, and difficulty with language comprehension. He had no history of surgery or trauma to the head and no neurological or otolaryngological disease. His mental status had previously been normal, and he had no complaints of headache, neck stiffness, fever, facial pain, otalgia, or otorhinorrhea. His medical history was remarkable for noninsulin-dependent diabetes mellitus and a cerebrovascular accident without clinical sequelae.

A nonenhanced computerized tomography (CT) scan of the head clearly revealed intraparenchymal air in the left temporal lobe (Fig. 1a). The sella turcica, sinuses, and mastoid appeared normal. A thorough otolaryngological examination including flexible fiberoptic nasopharyngoscopy was unremarkable.

Examination. The patient was referred to our institution 2 weeks after the onset of symptoms. He presented in a confused state, with a Wernicke's aphasia. A CT scan revealed intraventricular air, a low-density
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**Fig. 1.** Computerized tomography (CT) scans of the head. a: Initial nonenhanced axial CT scan showing intra-axial air density in the left temporal lobe, which from its spherical shape appears to be under tension. Subarachnoid air is also seen in the interpeduncular cistern. No evidence of the patient's previous cerebrovascular accident is noted. b and c: Axial (b) and coronal (c) scans obtained upon referral, showing a low-density lesion with an adjacent air collection in the left temporal lobe. Again, air is seen in the basal cisterns (around the cavernous sinus). Intraventricular air is now present in the left temporal horn of the lateral ventricle. d: Axial CT scan obtained after continued deterioration of the patient's mental status, showing large amounts of intraventricular air in the anterior horn of the lateral ventricles. Also note subarachnoid air in the right sylvian fissure and, more inferiorly, mass effect on the left temporal lobe.

left temporal lobe lesion with an adjacent air collection, and bone erosion of the left petrous pyramid (Fig. 1b and c). Computerized tomography of the temporal fossa verified a left petrous ridge defect (Fig. 2). The patient's mental status continued to deteriorate. Follow-up CT scans revealed increasing pneumocephalus with mass effect (Fig. 1d). The likelihood of tension pneumocephalus with intraventricular rupture justified aggressive surgical intervention.

**Operation.** The left middle fossa was approached via a wide left temporal craniotomy. The bone plate was separated from the dura, and bone was removed down to the middle fossa floor. Because of the likely presence of an intracranial lesion, the dura was opened widely. A multiseptated cystic lesion of the left temporal lobe with surrounding hypoechoic parenchyma was localized by means of intraoperative ultrasonography. Clear fluid was aspirated from the cystic lesion and multiple biopsy specimens were taken from what appeared to be the cyst wall and the adjacent hypoechoic temporal lobe. No tumor was seen on frozen section. The cyst was entered and noted to have a smooth lining. A communication with the ventricular system was apparent. The temporal lobe was then elevated off the floor of the middle fossa and a 5-mm encephalocele was seen entering the lateral aspect of the petrous ridge anteriorly. The encephalocele was amputated at the pial surface and removed from a smooth circular bone defect on the anterior lateral aspect of the petrous ridge. This defect had the appearance of an enlarged apical air cell. An additional 1-mm bone defect was seen in close proximity to the first, but contained no encephalocele. This defect was plugged with a piece of temporalis muscle overlaid with a strut of cortical bone. A free piece of pericranial graft was placed over the strut and sutured to the surrounding dura. Permanent sections of the biopsy specimens revealed no definite tumor, although the white matter surrounding the cyst was gliotic (Fig. 3). The specimen thought to be cyst wall was simply gliotic brain.

**Postoperative Course.** Postoperatively, the patient's condition was unchanged from his admission status. Serial brow-up skull films and a follow-up CT scan showed complete resolution of the intracranial air, with no evidence of residual cyst. Several months later, the patient died from pneumonia. His mental status and language comprehension had reportedly improved prior to his death. A request for autopsy was denied.
Discussion

Only 5% of all encephaloceles occur in the cranial base but, among these, the middle fossa is the most common site.6,17 Less than 10 cases of "spontaneous" encephaloceles of the middle fossa have been reported, and none has led to tension pneumocephalus due to rupture. Two major classes of spontaneous encephaloceles have been defined: congenital and acquired. Congenital encephaloceles refer to those arising in patients who present early in life and who have either a history of birth trauma or cranial-base abnormalities found incidentally during intracranial surgery for other problems. The class of acquired encephaloceles is further subdivided into: 1) traumatic, defined as those that present after obvious head trauma (injury or surgery);17,18 and 2) nontraumatic, referring to those that present spontaneously later in life without a history of CSF leak or obvious predisposing factors. This latter group suffers the consequences of "internal physiological trauma" over many years.

The pathogenesis of acquired nontraumatic encephaloceles can be explained as a consequence of normal anatamical and physiological factors occurring simultaneously in the right combinations and has been the subject of previous discussion.12,17 It is not uncommon to find multiple cranial defects, as in our case, of variable size and resembling pitholes in the middle fossa, particularly in the anteromedial aspect.12,21 These defects probably develop in the following way. Cerebrospinal fluid pulsatile/pressure dynamics unique to the middle fossa alter the bone architecture and cause a variable thinning of the superficial fossa floor to the point of eventual bone dehiscence of "critically thin areas" into the underlying air cells of the normally pneumatized skull base.12,13,21 The resulting bone excavations fill with cortical tissue and/or meninges, producing a meningocele or meningoencephalocele. Reflecting the dura from the middle fossa reveals that these "celes" separate cleanly from underlying bone and appear as nodules of growth upon the external surface of the brain.3 Interestingly, Russell21 reported that the dura appears normally to be thin and fenestrated in this region. Thus, it is not difficult to imagine that a sudden brisk rise in ICP from perhaps a cough or sneeze could cause rupture of an encephalocele through the thin dura, resulting in a CSF fistula. It was originally thought that trauma was a necessary antecedent to rupture, but several authors have presented autopsy cases supporting spontaneous rupture.14,15 Logically, signs and symptoms of CSF leak follow rupture, although in 30% of cases with a normal CSF pressure the leak abates spontaneously.15

Air can enter the cranial cavity through potentially patent fistulous tracts whenever extracranial pressure exceeds ICP, resulting in pneumocephalus. When the intracranial contents act as a one-way valve, tension pneumocephalus can occur as air enters but is unable to escape from the intracranial compartment. A localized tension pneumocephalus exerting sufficient mass effect could conceivably cause focal neurological deficits. If intraparenchymal dissection to the ventricular wall occurs, rupture into the ventricle may follow. Although the ICP in our patient was never actually recorded, it appeared that the pneumocephalus was under tension; this was based on a correlation of the patient's progressive clinical course with the appearance of the encephalocele on serial CT scans.

The dramatic spontaneous appearance of intracranial air should raise the question of a ruptured encephalocele, possibly of the middle fossa, even in patients who are atypically symptomatic. Tension pneumocephalus with rapid deterioration of mental status may ensue and should be anticipated so that timely surgical intervention in this life-threatening situation may proceed.

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